INFUNDIBULOMA : REPORT OF A CASE

AUTHOR(S):

ISHII, SHOZO; KANEDA, KAZUO

CITATION:

ISHII, SHOZO ...[et al]. INFUNDIBULOMA : REPORT OF A CASE. 日本外科宝函 1959, 28(5): 1903-1910

ISSUE DATE:

1959-06-01

URL:

http://hdl.handle.net/2433/206879

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INFUNDIBULOMA
REPORT OF A CASE

by

Shozo Ishii and Kazuo Kaneda

From the lst Surgical Division, Kyoto University Medical School
(Director : Prof. Dr. Chisato Araki)
Received for publication on May 31, 1959

INTRODUCTION

The purpose of this paper is to report a case of rare tumor in the region of the third ventricle. Though the histological structures of this tumor were entirely unfamiliar to us, we felt that they bore a close resemblance to those of infundibuloma which was first studied and described by Globus. We, however, still hesitated to report this as the seventh case of infundibuloma recorded in the literature since the first Globus's report[4] in 1942, and classified it provisionally as an astrocytic glioma.

Several months ago Dr. Bucy came to Japan and examined the sections of this tumor. His conclusion was that this tumor arose from pituicytes which had been named by him in 1932 to the cellular element of the neurohypophysis and the infundibular stalk. He also agreed us to put this tumor into the category of infundibuloma.

REPORT OF A CASE

History: R. K. A five-year-old girl, was admitted to the Kyoto University Hospital on July 7, 1955. She had been in reasonably good health until a month prior to the first admission to the other University Hospital in October 1954, when she commenced vomiting and impairment of the vision. She was operated upon there, and a cystic tumor was disclosed medially to the tip of the right temporal lobe which was partially removed. The histological diagnosis made by the former operater was astrocytoma. After the operation, she was getting along fairly well for abouf 5 months, then she again developed nausea and vomiting. Though the symptomatic treatment was started by her family doctor, her general conditions got progressively worse. A week before entering our hospital she became semicomatose.

Examination: The patient was semicomatose and emaciated. She was in the typical posture of "decebrate rigidity" with flexed upper extremities on the chest and extended legs. She had an obviously enlarged head, particularly a localized bulging of the bone around the former operative wound. She had a nystagmoid movement of eyes and the deviated visual axis toward left. There was bilateral optic atrophy. Both pupils reacted very poorly to light. The deep tendon reflexes were normal and no pathological reflex was found. A lumbar puncture yielded

cerebrospinal fluid under an initial pressure of 290mm of water.

Due to the poor cooperation of the patient other examinations could not be carried out.

Course: The first operation in our hospital was carried out on July 17, 1955. At the tip of the right temporal lobe, and beneath the thin layer of the cortex there was found a large cystic cavity. Since the cyst wall was smooth throughout, and the mural nodule was not visible, only an evacuation of the cyst content was carrid out. After the operation the child was still in coma and had to be fed by tube. Decerebrate rigidity continued and the bone flap was gradually bulged beneath the skin. On July 21, 1955, Torkildsen's operation was performed. The patient remained in the vegetative state after the second operation and died on November 7, 1955.

AUTOPSY

Gross findings: Towards the inferior surface of the right temporal lobe, there was a yellowish cortical area of degeneration with collapsed cyst wall at the site of the previous operation. At the medial border of the cyst, there was a large nodular tumor of hen-egg size which was hard in consistency. (Fig. 1). It involved the tip of the right temporal lobe, occupied the entire interpeduncular space, projecting forewards under the basofrontal surface and backwards encroaching upon the pons. The optic chiasm was not visible because it was burried in the tumor mass. On sectioning the brain, the third ventricle was displaced upwards and was reduced to a narrow slit. Tumor involved both hypothalamus and infundibular area, and extended posteriorly to the inferior surface of the pons. (Fig. 2).

Microscopic description: Tumor cells were predominantly elongated, unipolar or bipolar and few were triangular or astrocyte-like. (Fig. 3). They were quite similar to the cells normally found in the infundibulum or the posterior pituitary, the pituicytes. (Fig. 4). The nuclei of the fusiform cells were oval or elongated with moderate amount of chromatinic substance. The cytoplasmic processes were fairly thick and long, giving a "wiry" appearance. The tapering processes were sometimes aggregated into the streaming fibrous bundles in various directions. (Fig. 5). Around the blood vessels these processes were irregularly undulated and formed the fibrous meshwork. (Fig. 6). Throughout the tumor there were numerous vessels of irregular shape. In some, a fairly remarkable proliferation of the adventitial layer was recognized, but it was rather rare. Almost all blood vessels were lined with a very thin or membranous layer of endothelium. (Fig. 7), exhibiting features of sinusoid or vascular lakes. These features were again quite similar to those of the normal hypophysio-portal vessels in the infundibulum or the tuber cinereum. (Fig. 8). Throughout the tumor there were seen many bodies which stained homogeneously with eosin, colloidal droplets.

No mitosis, hemorrage, necrosis or other marked anaplastic features were observable. With various special stains, such as MAROLEY'S P. T. A. H. stain, CAJAL'S reduced silver stain or Bilschowski's stain for axis cylinder, neither typical glial elements nor nerve cells could be found.

DISCUSSION

A peculiar type of tumor in the infundibular region of the third ventricle was first studied and described by GLOBUS in 1942.[4] He reported two cases. One was a 13 and half-year-old boy and the other was a girl, aged 5 years. Both died after surgery. Necropsies revealed tumors of the almost similar character in the region around the third ventricle in either case. In microscopic structures, although two tumors presented few minor differences, both demonstrated similar morphorogical characteristics as will be described later. Thus, to this newly recognized tumor, the term "infundibuloma" was given by GLOBUS, for it indicated the origin and histological character of the tumor.

In 1947, PAPEZ and ECKER[6] reported a case of infundibuloma associated with precocious puberty in a boy, age 8. In their paper, a more emphasis was put in the determination of the correlation between the hypothalamic structure destroyed or spared by the tumor of the third ventricle and the occurrence of the precocious puberty than in the histological study of the tumor. However, the microscopic structures were similar to those described by GLOBUS, and some of the sections were in fact examined by GLOBUS who confirmed the diagnosis. FINE and GOLDFARB[3] descrberd a case of this tumor in a 6-year-old boy. POSENSER, MITCHENER and SKWAROK[8] described another one case of infundibuloma in a boy, age 1, who was the youngest of the patients that have so far been reported. Authors considered that the tumor was present since birth.

More recently, LISS and KAHN[5] reported a primary tumor of the posterior lobe of the hypophysis under the name of pituicytoma. The age of the patient was 47 years. GLOBUS advocated that infundibuloma was more likely to be encountered among the brain tumors in childhood, and in fact, all hitherto reported cases of infundibuloma were among the ages of younger generation except the last case. Also histological structures of the last case was somewhat different from those of our case and also from those of tumors described by GLOBUS. Thus, we were not quite certain whether the tumor which was reported by Liss and Kahn should be included in the category of infundibuloma or not. We, however, should like to include this, because it was considered that the tumor reported by them was also of neurohypophysial derivation.

Now, in order to place a given tumor in the category of infundibuloma, or to differentiate that from the tumors which bear some resembrance to infundibuloma, enough knowledge on the peculiarities or characteristis of this rare tumor is required.

Cellular element:

As was described by GLOBUS the predominant cell in infundibuloma is pituicytes. The name "pituicyte" was first given by BUCY[1] to the cell in the pars nervosa of the hypophysis. According to his description, pituicytes are of various shapes, but the majority of the cells tend to be either bipolar or unipolar. The bipolar cells usually have two very long processes which extend for long distance. And these processes are much longer than those of glial cells seen elsewhere. This is especially true of the infundibulum and stalk where the cells lie paralleled to the nerve fibers

passing into the infundibular process. The other cells are very irregular in shape and almost no two of these are alike. Pituicytes are unlike any cells of the central nervous system so far described and was considered that they constitute a new and special group of glial cells. These cells and their processes are most clearly seen in sections prepared according to the specific technique for either oligodendroglia or microglia. Using various stains for glia, Bucy could not find astrocytes anywhere in the pars nervosa.

More recently Liss and Kahn[5] studied on the morphology of pituicytes. According to them, pituicytes can be divided into four groups (1) bipolar (2) astrocyte-like (3) triangular and (4) glomerular. The bipolar pituicytes have elongated or oval cell bodies and their protoplasm may be either homogeneous (group 1A) or granular (group 1B). The pituicytes are capable of producing tumors which are formed by several of the above-named groups of pituicytes or by one of the groups.

They reported a case of pituicytoma formed predominantly by bipolar pituicytes with finely granular cytoplasm (group 1B). But all other infundibulomas reported, including our case, are different from their case in that they are mainly composed of bipolar pituicytes with homogeneous cytoplasms and very long tapering processes. In fact, the bipolar pituicytes with granular cytoplasm are found in normal neurohypophysis in small numbers. Thus, it can be assumed that in the formation of the tumor, infundibuloma, these cells may take a smaller part than that of the bipolar pituicytes with homogeneous cytoplasm.

Blood vessels:

As was described before, the blood vessels seen in the infundibuloma are quite unusual and peculiar. These vessels branch out irregularly, and appear as sinuses lined by a single layer of endothelial cell, which in turn are surrounded by a wide zone of a meshwork of fibers. For these, Globus gave the name "vascular lake". These characteristics of the blood vessels in tumor, however, are quite similar to those seen in the normal infundibular region. Popa and Fieldings[7], in 1930, found a system of blood vessels of a very unique appearance, in the normal tuber cinereum and gave a term of hypophysio-portal system. According to their description, these veins take origin from the sinusoids of the buccal portion of the hypophysis and from the capillaries of its neural portion, and ascend through the stalk to the region of the floor of the infundibular recess of the third ventricle where they break up into a secondary capillary net. In their course they acquire "glial sleaves" as far as the floor of the infundibular recess.

More recently, Christ,[2] studying the anatomy of the tuber cinereum, described on the structure of the hypophysial portal system. According to him, some branches of the superior hypophysial arteries pass downwards in the pars infundibulalis, giving rise in their early course to a mantle capillary plexus from which capillary loops extend into the infundibulum and infundibular stem. In its further course downwards, the capillary plexus becomes continuous with the sinusoid which in turn becomes connected with the sinusoid of the anterior lobe.

At any event, these vessels constitute a definite anatomical pattern and serve

as a landmark in the infundibular region. Also the presence of these vessels in infundibuloma may strongly suggest its site of origin.

Colloidal bodies:

In almost all cases of infundibuloma so far reported, numerous colloidal bodies which stain fairly well with eosin, have been recognized among the cellular elements of the tumor tissue and in some of the vessel walls. These also bear a strong resemblance to the colloidal substance or bodies found in the normal neuro-hypophysial region.

The supraoptic and paraventricular nuclei have long been considered as "diencephalic glands" because of the presence of "colloid" droplets in the cytoplasm of their cells. It is now widely believed that these granules or droplets contain the neurosecretory material. These tend to accumulate at nerve endings around the blood vessels and to be found extracellularly in the hypothalamus and neurohypophysis.

All above mentioned characteristics of the tumor have to be borne in mind for the histological diagnosis of infundibuloma. Tumor groups in which infundibuloma may most likely be misplaced or misidentified, are neurofibroma and spongioblastoma polare.

Histological picture of neurofibroma reveals many similarities to those of infundibuloma. Both show interlacing of parallel bundles of cells and cytoplasmic processes. Neurofibroma is generally believed to be composed of lemmocytes which parallel to the axis cylinders and nourish the latter. Also pituicytes which are the constituent cells of the infundibuloma, are believed to support, protect and perhaps nourish the nerve fiber. Bucy also noticed that pituicytes have a tendency to be arranged with their long axis paralleled to the course of nerve fibers in the pars nervosa. It may be for this reason that these two show a striking similarity in the sections prepared with a routine stain. These two, however, are readily distinguishable with the stain of silver impregnation.

Spongioblastoma polare is a relatively slow growing glioma with a predilection for the brain stem and is especially common along the optic tract and in the pons. Some investigators such as ZÜLCH[9] considered that the infundibuloma was just a modified spongioblastoma polare. He claimed that it was not wise to give such kind of "lokalisatorisch" name to a morphorogically and biologically identical tumor. In spongioblastoma, however, it is always possible to find many astrocytes besides the predominant spongioblastic cells, while in infundibuloma no typical astrocyte is usually observable. Also, in the former, we can find neither the peculier vascular pattern nor colloidal bodies as can be seen in the latter.

Thus, it might be reasonable to put our tumor into a tumor group of distinct and rare entity, infundibuloma.

SUMMARY

A primary tumor, infundibuloma, in the region of the third ventricle was described which occurred in a five-year-old girl, who died after three times of surgery. The histological features of this tumor were quite similar to those described by

GLOBUS. Characterics of this tumor were discussed in comparison wite the constituents of the normal infundibulum and neurohypophysis.

We would like to express our gratitude to Dr. ARAKI and to Dr. BUCY who kindly examined sections, and gave us many valuable advices.

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和文抄録

インフンディブローマ

石井昌三・金田一男

私達は第Ⅲ脳室底に発生した巨大な塊状腫瘍を経験したが，組織学的検索の結果，これが現在までに6例の報告例を見るに過ぎないインフンディブローマであることを確めた．症例は5才の女子，入院約10ヵ月前他の病院で右側頭葉腫瘍の診断の下に手術を受けた．その後諸症状（頭痛，視力障害，意識混濁発作等）の再現を見たため，吾々の病院に入院，二度の手術（腫瘍嚢腫内容除去，トルキルトセン氏手術）の後死亡した．剖検の結果第Ⅲ脳室底部より発生し前頭葉底面，右側頭葉下面，大脳脚間をみたす鶏卵大の腫瘍が存在した．組織学的には脳下垂体後葉及び漏斗の構成細胞である pituicyte を母細胞としたインフンディブローマであつた．

腫瘍組織像と正常下垂体後葉及び漏斗の組織像とを対比しつゝ，その構成細胞，血管系，コロイド体等の類似点を述べた．

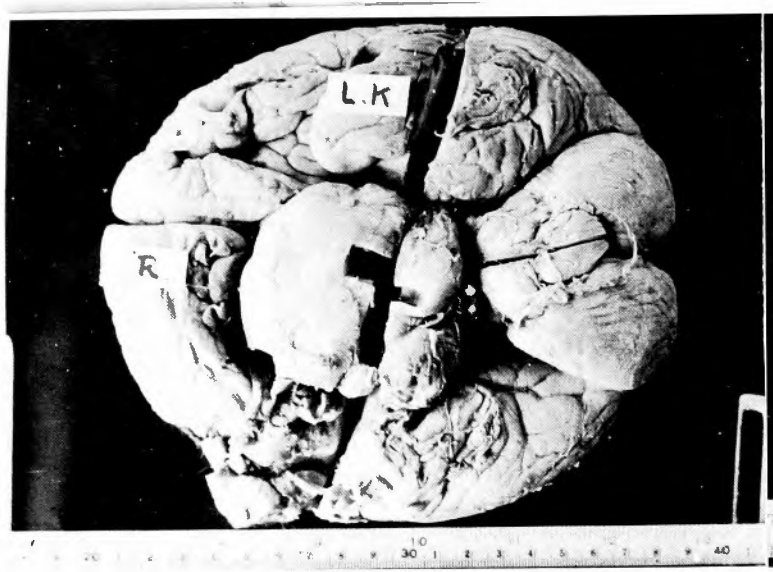

Fig. 1. Basal view of brain. Note huge centrally situated nodular tumor.

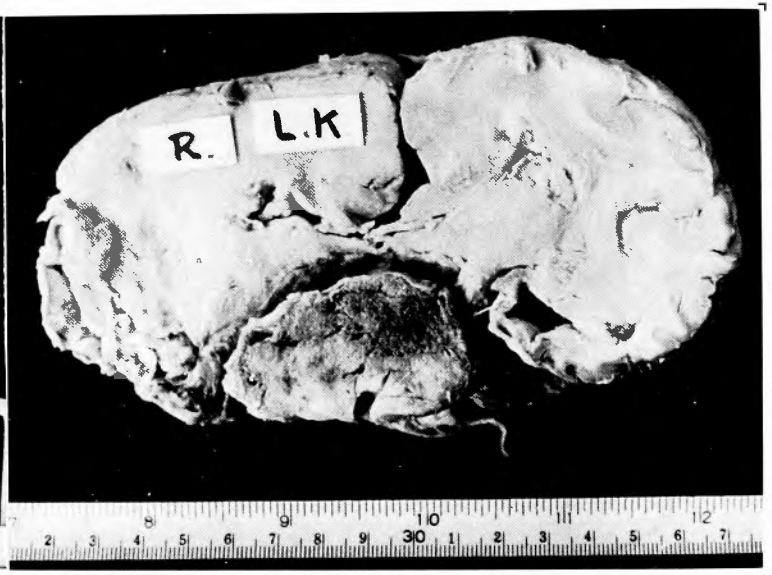

Fig. 2. Frontal Section of brain and tumor.

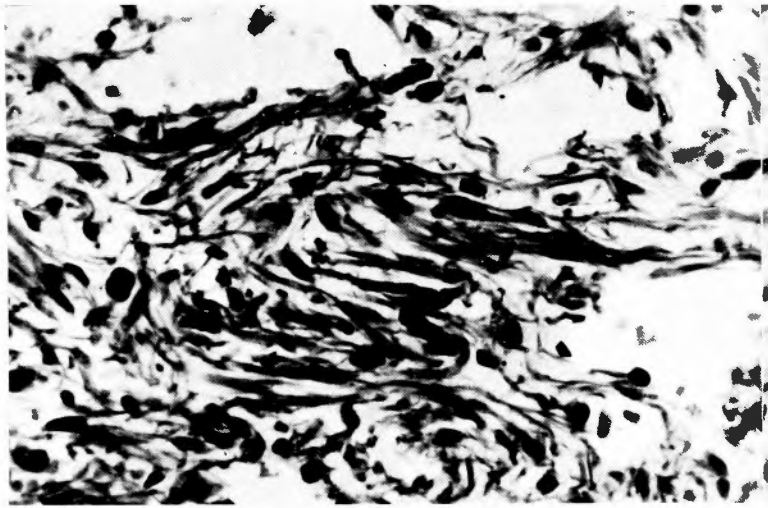

Fig. 3. Fusiform tumor cells. Note very long bipolar cytoplasmic processes and colloidal body. ×400 H. E. Stain

Fig. 4. Pituicytes in normal infundibular Stalk. ×400 H. E. Stain

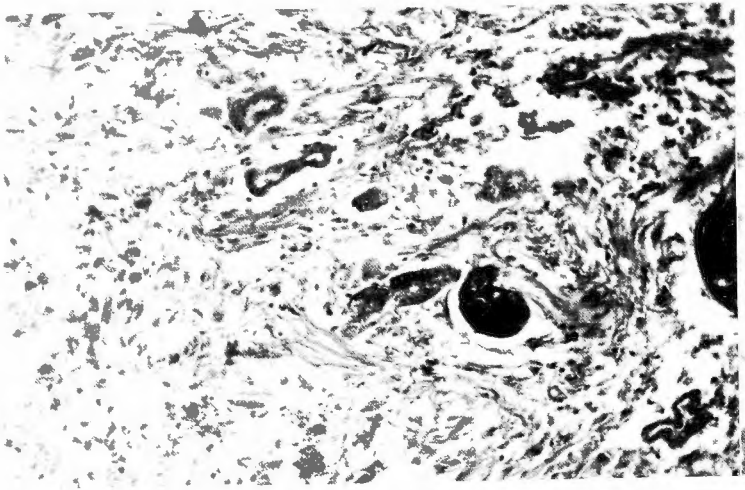

Fig. 5 Low magnification of tumor. Note "wiry' cytoplasmic processes which aggregated into the streaming bundles. ×200 H.E. Stain

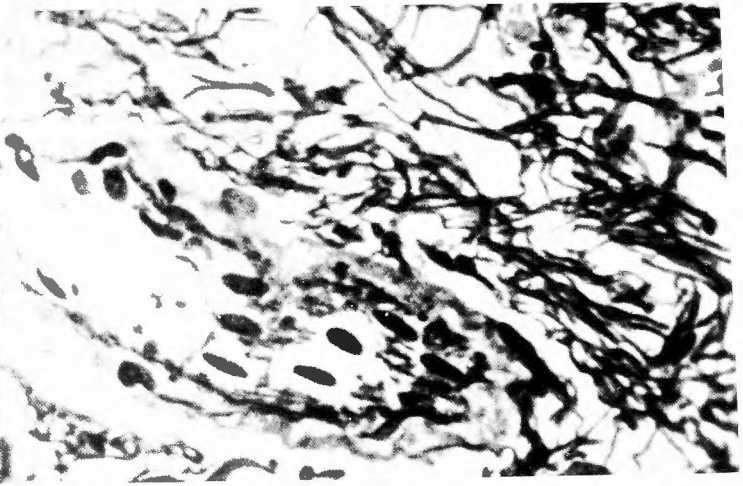

Fig. 6. Irregularly undulated cytoplasmic processes attached to the wall of blood vessel. ×400. reduced silber Stain.

Fig. 7. Sinusoid vessels of tumor. Blood vessels are lined with a very thin layer of endothelium. ×200. P T. A. H Stain.

Fig. 8. Blood vessels in normal infundibular stalk. ×200 P. T. A. H. Stain.